

- 
- Cardiology (290)
 - Clinical haematology/oncology (171)
 - Clinical pharmacology and toxicology
 - Clinical sciences (410)
 - Dermatology (131)
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 - Gastroenterology (183)
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 - Respiratory medicine (162)
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2600 MCQ with Illustrated Answer

Full Topics Note

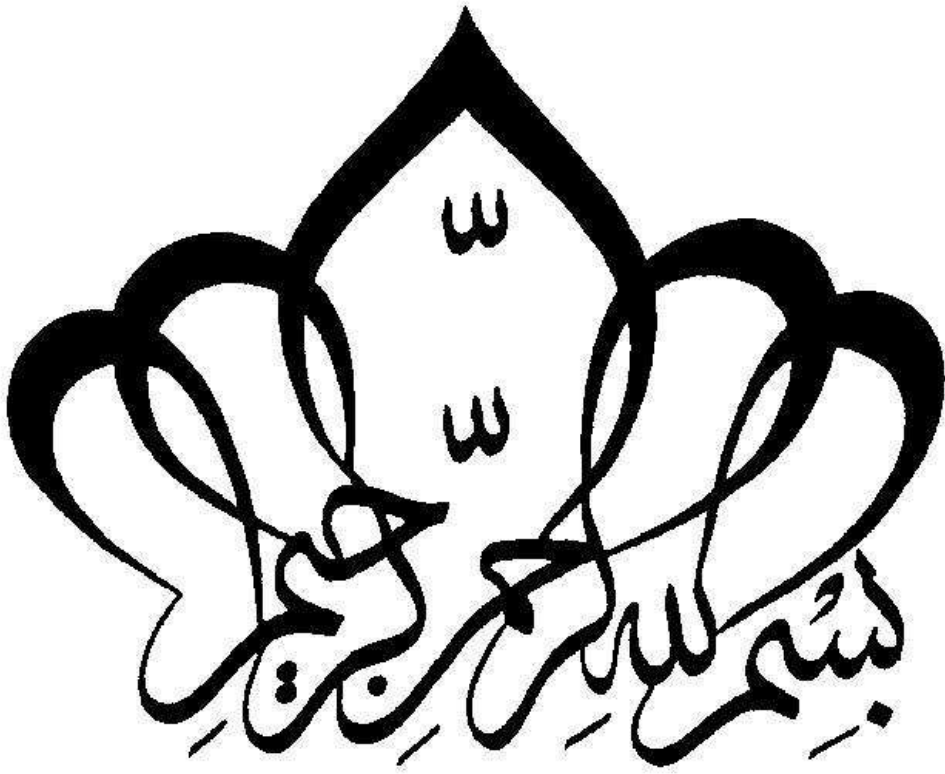
14 Chapter

Nephrology

P. M MCQ Bank 2017

VISIT...

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Caliente.COM



قَالُوا سُبْحَانَكَ لَا عِلْمَ لَنَا إِلَّا مَا عَلَّمْتَنَا
۞ إِنَّكَ أَنْتَ الْعَلِيمُ الْحَكِيمُ

إهداء

للكل آية من كتاب الله حفظناها ولكل معلومة قرأناها ونسيناها لعل الله يحمينا من النسيان ويروها إلينا
للكل شخص وعونا له في ظهر الغيب لعل الله أن يستجيب دعائنا
للكل مريض قرأ له ما أله لعل الله يشفيه ويعير إليه صحته وعافيته
للكل لحظة تألم بها أحببنا لعل الله لا يريهم الألم ولا يضييهم مرة أخرى
للكل موقف اجتجت به عيناى فلم تلبيانى لعل الله لا يضرنى بهما في حياتى
للكل حالم صاحب أهراف ومباوى لعل الله يحقق له مقاصده حتى يرضيه ويرضى عنه
للكل مجتهد لعل الله يجعل له من التوفيق أوفر حظ ونصيب ويهون له الأسباب كلها من حيث لا يدري من غير ضراء مضرة ولا فتنة مضلة
للكل من فقر شيئا في الحياة لعل الله يعيره إليه أو يعوضه بما يسعده
للكل شخص ليتعرفنا عنه لعل الله يجمعه بمن هم أفضل منا
وأخيرا وليس آخرا لأهلنا لعل الله يجعلها لهم صرقة جارية ترو إليهم قليلا من فضلهم علينا وتكون سببا يلم شملنا تحت سقف واحد بعد أن
هجرتنا الحروب والمصائب

يقول أحد الصالحين

إِذَا رَأَيْتَ أَنَّ اللَّهَ وَفَقَكَ لِإِخْوَةِ تَعْيْنِكَ عَلَى الْخَيْرِ.. فَأَعْلَمْ أَنَّ اللَّهَ يَرِيدُ بِكَ خَيْرًا !! وَإِذَا وَفَقَكَ اللَّهَ لِلدُّعَاءِ.. فَأَعْلَمْ أَنَّ اللَّهَ يَرِيدُ أَنْ يُعْطِيَكَ !! وَإِذَا
وَفَقَكَ لِقَضَاءِ حَوَائِجِ النَّاسِ فَأَعْلَمْ أَنَّ لَكَ يَتَخَلَّى عَنْكَ !! وَإِذَا وَفَقَكَ اللَّهَ لِذِكْرِهِ.. فَأَعْلَمْ أَنَّكَ مُحِبٌّ !!.. وَإِذَا أَحْبَبَّكَ أَحَدٌ.. وَنَصَرَكَ.. وَأَيَّرَكَ..
وَأُسْتَجَابَ لِدُعَائِكَ !!

This issue of **MRCPPART1 MADE EASY 2017** contains the following:

- Full topic note.
- Around 2600 MCQs from **PM 2017 BANK** covering 14 chapter in internal medicine
- External links for the related guidelines (such as NICE, SIGN, RCP ...etc).
- Comments with illustrative pictures and links to external media.
- Most Recommended QBank for MRCP Part 1, Arab Board, Jordanian Board and other internal medicine assessment exams.

Please Note that

Each chapter begins with notes section. These notes are repeated under the Questions in the website. In these collections the notes are not repeated under each question except in some to avoid unnecessary repetition. For clarity and convenience the title of the related note is only mentioned after the comments in case the reader want to refer back to the respective note in the notes section.

The illustration below is to show and Example

☐	Minimal change disease
☐	Post-streptococcal glomerulonephritis
☐	Goodpasture's syndrome

Goodpasture's syndrome

- IgG deposits on renal biopsy
- anti-GBM antibodies

• These changes are characteristic of Goodpasture's syndrome.

[Discuss and give feedback](#)

Goodpasture's syndrome 

Comment of the Q

title of the note in case the reader want to refer back to the notes sections

MRCPPART1 MADE EASY 2017

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إعداد وتنسيق

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&

A. MURAD

لا تنسونا من صالح دعائكم

ما كان من خطأ فمن أنفسنا وما كان من توفيق فمن الله

تم بحمد الله وتوفيقه ومنه

جعل الله عملنا متقبلاً خالصاً لوجهه الكريم

Reference ranges

Reference ranges vary according to individual labs. All values are for adults unless otherwise stated

Full blood count

Haemoglobin	Men: 13.5-18 g/dl Women: 11.5-16 g/dl
Mean cell volume	82-100 fl
Platelets	150-400 * 10 ⁹ /l
White blood cells	4-11 * 10 ⁹ /l

Urea and electrolytes

Sodium	135-145 mmol/l
Potassium	3.5 - 5.0 mmol/l
Urea	2.0-7 mmol/l
Creatinine	55-120 umol/l
Bicarbonate	22-28 mmol/l
Chloride	95-105 mmol/l

Liver function tests

Bilirubin	3-17 umol/l
Alanine transferase (ALT)	3-40 iu/l
Aspartate transaminase (AST)	3-30 iu/l
Alkaline phosphatase (ALP)	30-100 umol/l
Gamma glutamyl transferase (yGT)	8-60 u/l
Albumin	35-50 g/l
Total protein	60-80 g/l

Other haematology

Erythrocyte sedimentation rate (ESR)	Men: < (age / 2) mm/hr Women: < ((age + 10) / 2) mm/hr
Prothrombin time (PT)	10-14 secs
Activated partial thromboplastin time (APTT)	25-35 secs
Ferritin	20-230 ng/ml
Vitamin B12	200-900 ng/l
Folate	3.0 nmol/l
Reticulocytes	0.5-1.5%
D-Dimer	< 400 ng/ml

Other biochemistry

Calcium	2.1-2.6 mmol/l
Phosphate	0.8-1.4 mmol/l
CRP	< 10 mg/l
Thyroid stimulating hormone (TSH)	0.5-5.5 mu/l
Free thyroxine (T4)	9-18 pmol/l
Total thyroxine (T4)	70-140 nmol/l
Amylase	70-300 u/l

Uric acid	0.18-0.48 mmol/l
-----------	------------------

Arterial blood gases

pH	7.35 - 7.45
pCO2	4.5 - 6.0 kPa
pO2	10 - 14 kPa

Lipids

Desirable lipid values depend on other risk factors for cardiovascular disease, below is just a guide:

Total cholesterol	< 5 mmol/l
Triglycerides	< 2 mmol/l
HDL cholesterol	> 1 mmol/l
LDL cholesterol	< 3 mmol/l

Chapter: Nephrology

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Chapter: Nephrology

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Acute kidney injury: acute tubular necrosis vs. prerenal uraemia

Prerenal uraemia - kidneys hold on to sodium to preserve volume

	Pre-renal uraemia	Acute tubular necrosis
Urine sodium	< 20 mmol/L	> 30 mmol/L
Fractional sodium excretion*	< 1%	> 1%
Fractional urea excretion**	< 35%	>35%
Urine:plasma osmolality	> 1.5	< 1.1
Urine:plasma urea	> 10:1	< 8:1
Specific gravity	> 1020	< 1010
Urine	'bland' sediment	brown granular casts
Response to fluid challenge	Yes	No

*fractional sodium excretion = (urine sodium/plasma sodium) / (urine creatinine/plasma creatinine) x 100

**fractional urea excretion = (urine urea /blood urea) / (urine creatinine/plasma creatinine) x 100

External Links

[Royal College of Physicians](#)

2012 Recognising acute kidney injury

Acute vs. chronic renal failure

Best way to differentiate is renal ultrasound - most patients with CRF have bilateral small kidneys

Exceptions

- autosomal dominant polycystic kidney disease
- diabetic nephropathy
- amyloidosis
- HIV-associated nephropathy

Other features suggesting CRF rather than ARF:

- hypocalcaemia (due to lack of vitamin D)

ADPKD

Autosomal dominant polycystic kidney disease (ADPKD) is the most common inherited cause of kidney disease, affecting 1 in 1,000 Caucasians. Two disease loci have been identified, PKD1 and PKD2, which code for polycystin-1 and polycystin-2 respectively

ADPKD type 1	ADPKD type 2
85% of cases	15% of cases
Chromosome 16	Chromosome 4
Presents with renal failure earlier	

The screening investigation for relatives is abdominal ultrasound:

Ultrasound diagnostic criteria (in patients with positive family history):

- two cysts, unilateral or bilateral, if aged < 30 years
- two cysts in both kidneys if aged 30-59 years
- four cysts in both kidneys if aged > 60 years



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Extensive cysts are seen in an enlarged kidney

ADPKD: features

Features

- hypertension
- recurrent UTIs
- abdominal pain
- renal stones
- haematuria
- chronic kidney disease

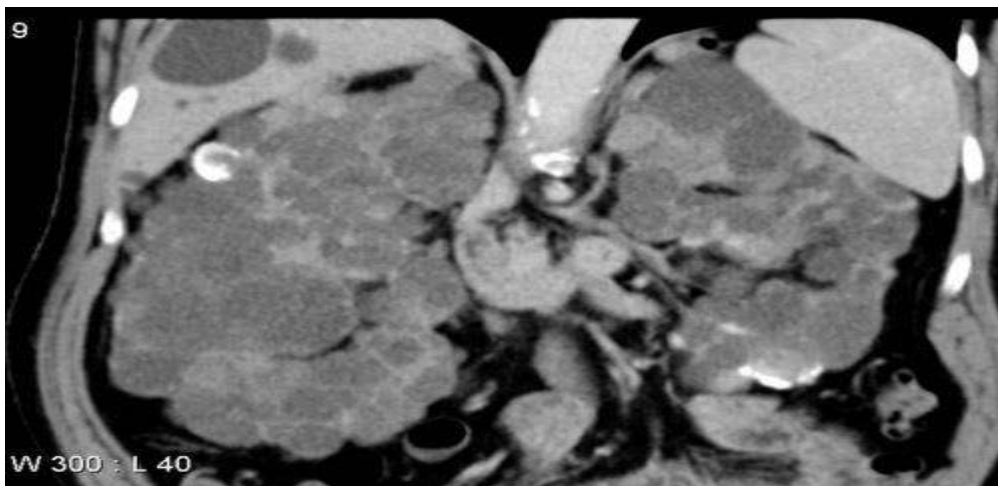
Extra-renal manifestations

- liver cysts (70%)
- berry aneurysms (8%)
- cardiovascular system: mitral valve prolapse, mitral/tricuspid incompetence, aortic root dilation, aortic dissection
- cysts in other organs: pancreas, spleen; very rarely: thyroid, oesophagus, ovary



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Extensive cysts are seen in an enlarged kidney



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CT of the abdomen demonstrates both kidneys to be markedly enlarged by innumerable cysts ranging in size from a few millimeters to multiple centimeters with cysts also present in the liver



© Image used on license from [Radiopaedia](https://radiopaedia.org/)



CT showing multiple cysts of varying sizes in the liver, and bilateral kidneys with little remaining normal renal parenchyma.

Alport's syndrome

♦ Alport's syndrome is usually inherited in an X-linked dominant pattern*. It is due to a defect in the gene which codes for type IV collagen resulting in an abnormal glomerular-basement membrane (GBM). The disease is more severe in males with females rarely developing renal failure

♦ A favourite question is an Alport's patient with a failing renal transplant. This may be caused by the presence of anti-GBM antibodies leading to a Goodpasture's syndrome like picture

♦Alport's syndrome usually presents in childhood. **The following features may be seen:**

- microscopic haematuria
- progressive renal failure
- bilateral sensorineural deafness
- lenticonus: protrusion of the lens surface into the anterior chamber
- retinitis pigmentosa
- renal biopsy: splitting of lamina densa seen on electron microscopy

*in around 85% of cases - 10-15% of cases are inherited in an autosomal recessive fashion with rare autosomal dominant variants existing

Amyloidosis

Overview

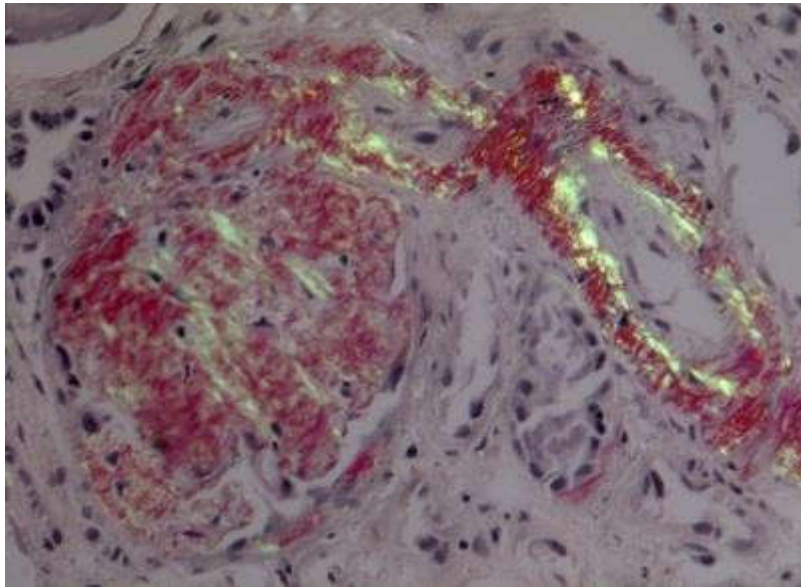
- amyloidosis is a term which describes the extracellular deposition of an insoluble fibrillar protein termed amyloid
- amyloid is derived from many different precursor proteins
- in addition to the fibrillar component, amyloid also contains a non-fibrillary protein called amyloid-P component, derived from the acute phase protein serum amyloid P
- other non-fibrillary components include apolipoprotein E and heparan sulphate proteoglycans
- the accumulation of amyloid fibrils leads to tissue/organ dysfunction

Classification

- systemic or localized
- further characterised by precursor protein (e.g. AL in myeloma - A for Amyloid, L for immunoglobulin Light chain fragments)

Diagnosis

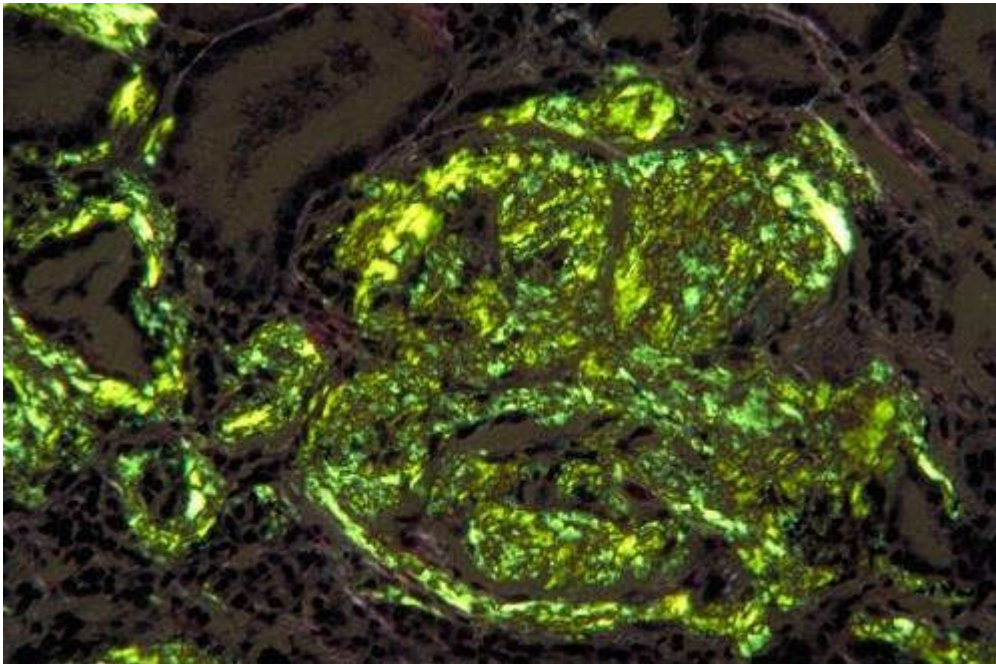
- Congo red staining: apple-green birefringence
- serum amyloid precursor (SAP) scan
- biopsy of rectal tissue



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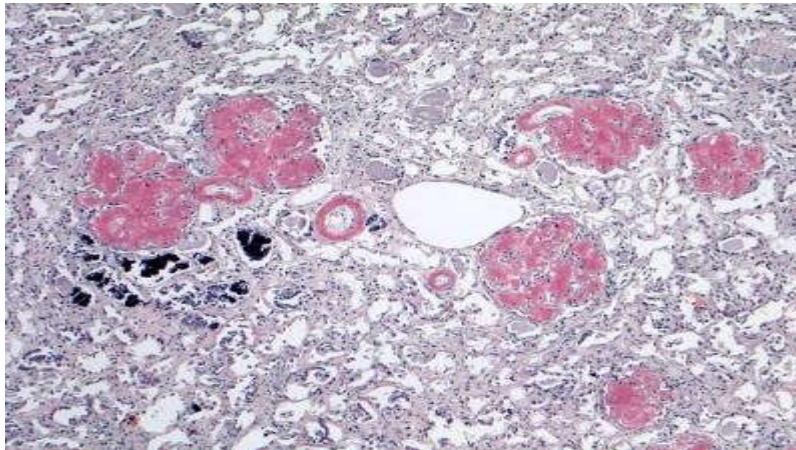
Renal amyloid with congo red staining - apple-green birefringence



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Renal amyloid with congo red staining - apple-green birefringence



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Congo red staining. Amyloid deposits are seen in both the arteries/arterioles and within the glomerulus. The deposit of amyloid within the mesangium is not dissimilar to the nodular lesions seen in diabetic nephropathy

Amyloidosis: types

AL amyloid:

- L for immunoglobulin Light chain fragment
- due to myeloma, Waldenstrom's, MGUS
- features include: cardiac and neurological involvement, macroglossia, periorbital ecchymoses

AA amyloid

- A for precursor serum amyloid A protein, an acute phase reactant
- seen in chronic infection/inflammation
- e.g. TB, bronchiectasis, rheumatoid arthritis
- features: renal involvement most common feature

Beta-2 microglobulin amyloidosis

- precursor protein is beta-2 microglobulin, part of the major histocompatibility complex
- associated with patients on renal dialysis

ARPKD

♦Autosomal recessive polycystic kidney disease (ARPKD) is much less common than autosomal dominant disease (ADPKD). It is due to a defect in a gene located on chromosome 6 which encodes fibrocystin, a protein important for normal renal tubule development.

♦**Diagnosis** may be made on prenatal ultrasound or in early infancy with abdominal masses and renal failure. Newborns may also have features consistent with Potter's syndrome secondary to oligohydramnios. End-stage renal failure develops in childhood. Patients also typically have liver involvement, for example portal and interlobular fibrosis.

♦**Renal biopsy** typically shows multiple cylindrical lesions at right angles to the cortical surface.

Cholesterol embolisation

Overview

- cholesterol emboli may break off causing renal disease
- seen more commonly in arteriopathies, abdominal aortic aneurysms

Features

- eosinophilia
 - purpura
 - renal failure
 - livedo reticularis
-

Chronic kidney disease: causes

Common causes of chronic kidney disease:l:

- diabetic nephropathy
- chronic glomerulonephritis
- chronic pyelonephritis
- hypertension
- adult polycystic kidney disease

Chronic kidney disease: eGFR and classification

Serum creatinine may not provide an accurate estimate of renal function due to differences in muscle. For this reason formulas were develop to help estimate the glomerular filtration rate (estimated GFR or eGFR). The most commonly used formula is the Modification of Diet in **Renal Disease (MDRD) equation, which uses the following variables:**

- serum creatinine
- age
- gender
- ethnicity

Factors which may affect the result

- pregnancy
- muscle mass (e.g. amputees, body-builders)
- eating red meat 12 hours prior to the sample being taken

CKD may be classified according to GFR:

CKD stage	GFR range
1	Greater than 90 ml/min, with some sign of kidney damage on other tests (if all the kidney tests* are normal, there is no CKD)
2	60-90 ml/min with some sign of kidney damage (if kidney tests* are normal, there is no CKD)
3a	45-59 ml/min, a moderate reduction in kidney function
3b	30-44 ml/min, a moderate reduction in kidney function
4	15-29 ml/min, a severe reduction in kidney function
5	Less than 15 ml/min, established kidney failure - dialysis or a kidney transplant may be needed

*i.e. normal U&Es and no proteinuria

External Links

[NICE](#)

2014 Chronic kidney disease guidelines

Chronic kidney disease: hypertension

♦The majority of patients with chronic kidney disease (CKD) will require more than two drugs to treat hypertension. ACE inhibitors are first line and are particularly helpful in proteinuric renal disease (e.g. diabetic nephropathy). As these drugs tend to reduce filtration pressure a small fall in glomerular filtration pressure (GFR) and rise in creatinine can be expected. NICE suggest that a decrease in eGFR of up to 25% or a rise in creatinine of up to 30% is acceptable, although any rise should prompt careful monitoring and exclusion of other causes (e.g. NSAIDs). A rise greater than this may indicate underlying renovascular disease.

♦Furosemide is useful as a anti-hypertensive in patients with CKD, particularly when the GFR falls to below 45 ml/min*. It has the added benefit of lowering serum potassium. High doses are usually required. If the patient becomes at risk of dehydration (e.g. Gastroenteritis) then consideration should be given to temporarily stopping the drug

*the NKF K/DOQI guidelines suggest a lower cut-off of less than 30 ml/min

External Links

[NICE](#)

2014 CKD guidelines

[NKF K/DOQI](#)

Use of diuretics in CKD

Chronic kidney disease: proteinuria

Proteinuria is an important marker of chronic kidney disease, especially for diabetic nephropathy. NICE recommend using the albumin:creatinine ratio (ACR) in preference to the protein:creatinine ratio (PCR) when identifying patients with proteinuria as it has greater sensitivity. For quantification and monitoring of proteinuria, PCR can be used as an alternative, although ACR is recommended in diabetics. Urine reagent strips are not recommended unless they express the result as an ACR

Approximate equivalent values

ACR (mg/mmol)	PCR (mg/mmol)	Urinary protein excretion (g/24 h)
30	50	0.5
70	100	1

Collecting an ACR sample:

- by collecting a 'spot' sample it avoids the need to collect urine over a 24 hour period in order to detect or quantify proteinuria
- should be a first-pass morning urine specimen
- if the initial ACR is between 3 mg/mmol and 70 mg/mmol, this should be confirmed by a subsequent early morning sample. If the initial ACR is 70 mg/mmol or more, a repeat sample need not be tested.

Interpreting the ACR results:

- the NICE guidelines state 'regard a confirmed ACR of 3 mg/mmol or more as clinically important proteinuria'

NICE recommendations for referral to a nephrologist:

- a urinary albumin:creatinine ratio (ACR) of 70 mg/mmol or more, unless known to be caused by diabetes and already appropriately treated
- a urinary ACR of 30 mg/mmol or more, together with persistent haematuria (two out of three dipstick tests show 1+ or more of blood) after exclusion of a urinary tract infection.
- consider referral to a nephrologist for people with an ACR between 3-29 mg/mmol who have persistent haematuria and other risk factors such as a declining eGFR, or cardiovascular disease.

Frequency of monitoring eGFR (number of times per year by eGFR and ACR categories) for people with or at risk of CKD:

eGFR categories (mL/min/1.73 m ²)	ACR categories (mg/mmol)		
	A1 (< 3) Normal to mildly increased	A2 (3-30) Moderately increased	A3 (> 30) Severely increased
G1 ≥90 Normal and high	≤ 1	1	≥ 1
G2 60-89 Mild reduction related to normal range for a young adult	≤ 1	1	≥ 1
G3a 45-59 Mild to moderate reduction	1	1	2
G3b 30-44 Moderate to severe reduction	≤ 2	2	≥ 2
G4 15-29 Severe reduction	2	2	3
G5 <15 Kidney failure	4	≥ 4	≥ 4

External Links

[NICE](#)

2014 Chronic kidney disease guidelines

[SIGN](#)

2008 Diagnosis and management of chronic kidney disease

Cytomegalovirus

Cytomegalovirus (CMV) is one of the herpes viruses. It is thought that around 50% of people have been exposed to the CMV virus although it only usually causes disease in the immunocompromised, for example people with HIV or those on immunosuppressants following organ transplantation.

Pathophysiology

- infected cells have a 'Owl's eye' appearance due to intranuclear inclusion bodies

Patterns of disease

Congenital CMV infection

- features include growth retardation, pinpoint petechial 'blueberry muffin' skin lesions, microcephaly, sensorineural deafness, encephalitis (seizures) and hepatosplenomegaly

CMV mononucleosis

- infectious mononucleosis-like illness
- may develop in immunocompetent individuals

CMV retinitis

- common in HIV patients with a low CD4 count (< 50)
- presents with visual impairment e.g. 'blurred vision'. Fundoscopy shows retinal haemorrhages and necrosis, often called 'pizza' retina
- IV ganciclovir is the treatment of choice



Fundus photograph showing CMV retinitis. Credit: National Eye Institute, National Institutes of Health

CMV encephalopathy

- seen in patients with HIV who have low CD4 counts

CMV pneumonitis

CMV colitis

Diabetic nephropathy: stages

Diabetic nephropathy may be classified as occurring in five stages*:

Stage 1

- hyperfiltration: increase in GFR
- may be reversible

Stage 2 (silent or latent phase)

- most patients do not develop microalbuminuria for 10 years
- GFR remains elevated

Stage 3 (incipient nephropathy)

- microalbuminuria (albumin excretion of 30 - 300 mg/day, dipstick negative)

Stage 4 (overt nephropathy)

- persistent proteinuria (albumin excretion > 300 mg/day, dipstick positive)
- hypertension is present in most patients
- histology shows diffuse glomerulosclerosis and focal glomerulosclerosis (Kimmelstiel-Wilson nodules)

Stage 5

- end-stage renal disease, GFR typically < 10ml/min
- renal replacement therapy needed

The timeline given here is for type 1 diabetics. Patients with type 2 diabetes mellitus (T2DM) progress through similar stages but in a different timescale - some T2DM patients may progress quickly to the later stages

External Links

[SIGN](#)

Diabetic nephropathy guidelines

Erythropoietin

Erythropoietin is a haematopoietic growth factor that stimulates the production of erythrocytes. The main uses of erythropoietin are to treat the anaemia associated with chronic kidney disease and that associated with cytotoxic therapy.

Side-effects of erythropoietin

- accelerated hypertension potentially leading to encephalopathy and seizures (blood pressure increases in 25% of patients)
- bone aches
- flu-like symptoms
- skin rashes, urticaria
- pure red cell aplasia* (due to antibodies against erythropoietin)
- raised PCV increases risk of thrombosis (e.g. Fistula)
- iron deficiency 2nd to increased erythropoiesis

There are a number of reasons why patients may fail to respond to erythropoietin therapy:

- iron deficiency
- inadequate dose
- concurrent infection/inflammation
- hyperparathyroid bone disease
- aluminium toxicity

*the risk is greatly reduced with darbepoetin

External Links

[Postgraduate Medical Journal](#)

Review of erythropoietin

Fanconi syndrome

Fanconi syndrome describes a generalised disorder of renal tubular transport in the proximal convoluted tubule resulting in:

- type 2 (proximal) renal tubular acidosis
- polyuria
- aminoaciduria
- glycosuria
- phosphaturia
- osteomalacia

Causes

- cystinosis (most common cause in children)
 - Sjogren's syndrome
 - multiple myeloma
 - nephrotic syndrome
 - Wilson's disease
-

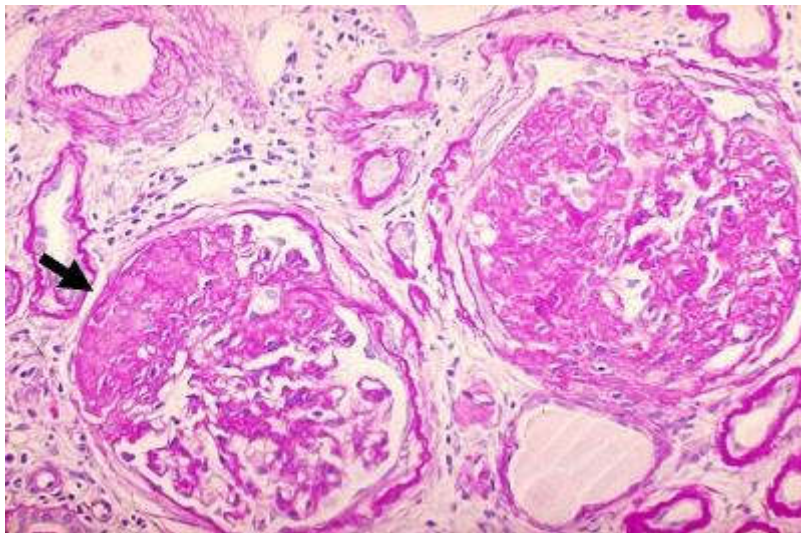
Focal segmental glomerulosclerosis

Focal segmental glomerulosclerosis is cause of nephrotic syndrome and chronic kidney disease. It generally presents in young adults.

Causes

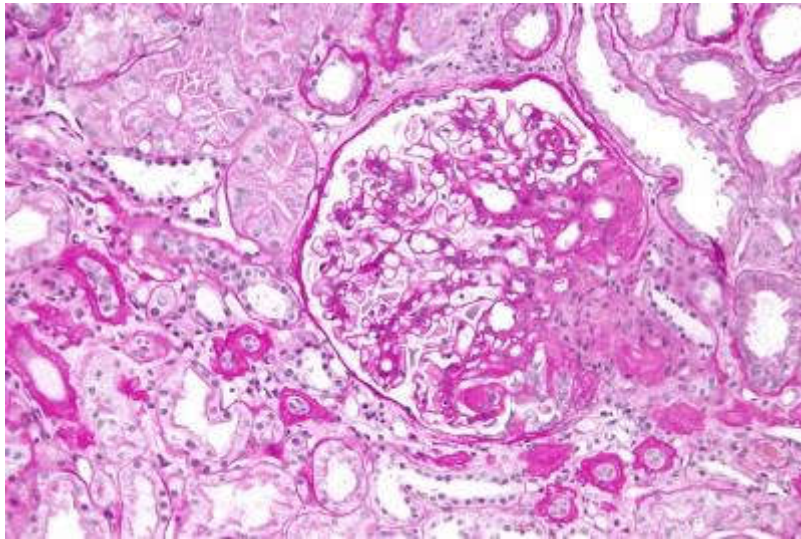
- idiopathic
- secondary to other renal pathology e.g. IgA nephropathy, reflux nephropathy
- HIV
- heroin
- Alport's syndrome
- sickle-cell

Focal segmental glomerulosclerosis is noted for having a high recurrence rate in renal transplants.



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Sclerosis of the glomerulus is seen next to Bowman's capsule



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Sclerosis is seen in the perihilar region of the glomerulus

Glomerulonephritides

Knowing a few key facts is the best way to approach the difficult subject of glomerulonephritis:

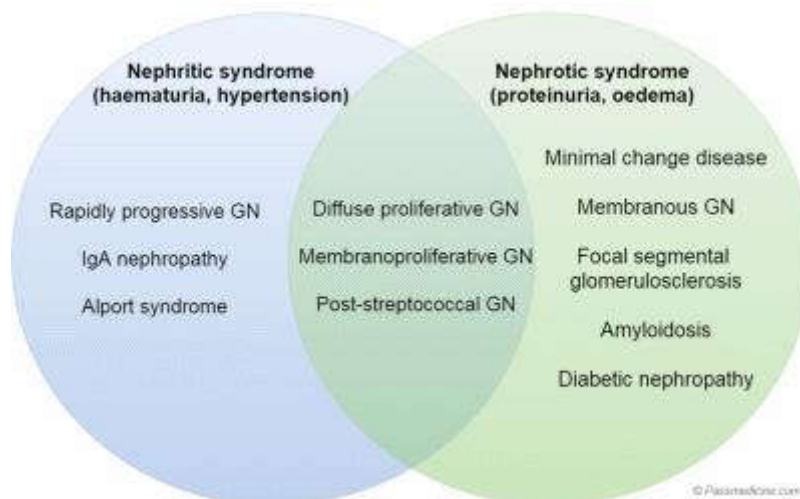


Diagram showing the glomerulonephritides and how they typically present

Typically presents with nephritic syndrome (haematuria, hypertension)

Rapidly progressive glomerulonephritis - aka crescentic glomerulonephritis

- rapid onset, often presenting as acute kidney injury
- causes include Goodpasture's, ANCA positive vasculitis

IgA nephropathy - aka Berger's disease, mesangioproliferative GN

- typically young adult with haematuria following an URTI

Mixed nephritic/nephrotic presentation

Diffuse proliferative glomerulonephritis

- classical post-streptococcal glomerulonephritis in child
- presents as nephritic syndrome / acute kidney injury
- most common form of renal disease in SLE

Membranoproliferative glomerulonephritis (mesangiocapillary)

- type 1: cryoglobulinaemia, hepatitis C
- type 2: partial lipodystrophy

Typically presents with nephrotic syndrome (proteinuria, oedema)

Minimal change disease

- typically a child with nephrotic syndrome (accounts for 80%)
- causes: Hodgkin's, NSAIDs
- good response to steroids

Membranous glomerulonephritis

- presentation: proteinuria / nephrotic syndrome / chronic kidney disease
- cause: infections, rheumatoid drugs, malignancy
- 1/3 resolve, 1/3 respond to cytotoxics, 1/3 develop chronic kidney disease

Focal segmental glomerulosclerosis

- may be idiopathic or secondary to HIV, heroin
- presentation: proteinuria / nephrotic syndrome / chronic kidney disease

External Links

[Postgraduate Medical Journal](#)

Acute glomerulonephritis

Glomerulonephritis and low complement

Disorders associated with glomerulonephritis and low serum complement levels

- post-streptococcal glomerulonephritis
- subacute bacterial endocarditis
- systemic lupus erythematosus
- mesangiocapillary glomerulonephritis

External Links

[Postgraduate Medical Journal](#)

Acute glomerulonephritis

Goodpasture's syndrome

Goodpasture's syndrome is rare condition associated with both pulmonary haemorrhage and rapidly progressive glomerulonephritis. It is caused by anti-glomerular basement membrane (anti-GBM) antibodies against type IV collagen. Goodpasture's syndrome is more common in men (sex ratio 2:1) and has a bimodal age distribution (peaks in 20-30 and 60-70 age bracket). It is associated with HLA DR2.

Features

- pulmonary haemorrhage
- followed by rapidly progressive glomerulonephritis

Factors which increase likelihood of pulmonary haemorrhage:

- smoking
- lower respiratory tract infection
- pulmonary oedema
- inhalation of hydrocarbons
- young males

Investigations:

- renal biopsy: linear IgG deposits along basement membrane
- raised transfer factor secondary to pulmonary haemorrhages

Management:

- plasma exchange (plasmapheresis)
 - steroids
 - cyclophosphamide
-

Haematuria

♦The management of patients with haematuria is often difficult due to the absence of widely followed guidelines. It is sometimes unclear whether patients are best managed in primary care, by urologists or by nephrologists.

♦The terminology surrounding haematuria is changing. Microscopic or dipstick positive haematuria is increasingly termed non-visible haematuria whilst macroscopic haematuria is termed visible haematuria. Non-visible haematuria is found in around 2.5% of the population.

Causes of transient or spurious non-visible haematuria

- urinary tract infection
- menstruation
- vigorous exercise (this normally settles after around 3 days)
- sexual intercourse

Causes of persistent non-visible haematuria

- cancer (bladder, renal, prostate)
- stones
- benign prostatic hyperplasia
- prostatitis
- urethritis e.g. *Chlamydia*
- renal causes: IgA nephropathy, thin basement membrane disease

Spurious causes - red/orange urine, where blood is not present on dipstick

- foods: beetroot, rhubarb
- drugs: rifampicin, doxorubicin.

Chapter: Nephrology

Management

Current evidence does not support screening for haematuria. The incidence of non-visible haematuria is similar in patients taking aspirin/warfarin to the general population hence these patients should also be investigated.

Testing

- urine dipstick is the test of choice for detecting haematuria
- persistent non-visible haematuria is often defined as blood being present in 2 out of 3 samples tested 2-3 weeks apart
- renal function, albumin:creatinine (ACR) or protein:creatinine ratio (PCR) and blood pressure should also be checked
- urine microscopy may be used but time to analysis significantly affects the number of red blood cells detected

NICE urgent cancer referral guidelines were updated in 2015.

Urgent referral (i.e. within 2 weeks)

Aged ≥ 45 years AND:

- unexplained visible haematuria without urinary tract infection, or
- visible haematuria that persists or recurs after successful treatment of urinary tract infection

Aged ≥ 60 years AND have unexplained nonvisible haematuria and either dysuria or a raised white cell count on a blood test

Non-urgent referral

Aged ≥ 60 years with recurrent or persistent unexplained urinary tract infection

Since the investigation (or not) of non-visible haematuria is such a common dilemma a number of guidelines have been published. **They generally agree with NICE guidance, of note:**

- patients under the age of 40 years with normal renal function, no proteinuria and who are normotensive do not need to be referred and may be managed in primary care

External Links

[NICE](#)

2015 Suspected cancer: recognition and referral

Haemolytic uraemic syndrome

Haemolytic uraemic syndrome is generally seen in young children and produces a triad of:

- acute renal failure
- microangiopathic haemolytic anaemia
- thrombocytopenia

Causes

- post-dysentery - classically E coli 0157:H7 ('verotoxigenic', 'enterohaemorrhagic')
- tumours
- pregnancy
- ciclosporin, the Pill
- systemic lupus erythematosus
- HIV

Investigations

- full blood count: anaemia, thrombocytopenia, fragmented blood film
- U&E: acute renal failure
- stool culture

Management

- treatment is supportive e.g. Fluids, blood transfusion and dialysis if required
- there is no role for antibiotics, despite the preceding diarrhoeal illness in many patients
- the indications for plasma exchange in HUS are complicated. As a general rule plasma exchange is reserved for severe cases of HUS not associated with diarrhoea

External Links

Renal.org

2009 atypical haemolytic uraemic syndrome guidelines

British Journal of Haematology

2012 TTP guidelines

Henoch-Schonlein purpura

Henoch-Schonlein purpura (HSP) is an IgA mediated small vessel vasculitis. There is a degree of overlap with IgA nephropathy (Berger's disease). HSP is usually seen in children following an infection.

Features:

- palpable purpuric rash (with localized oedema) over buttocks and extensor surfaces of arms and legs
- abdominal pain
- polyarthrititis
- features of IgA nephropathy may occur e.g. haematuria, renal failure



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Treatment

- analgesia for arthralgia
- treatment of nephropathy is generally supportive. There is inconsistent evidence for the use of steroids and immunosuppressants

Prognosis

- usually excellent, HSP is a self-limiting condition, especially in children without renal involvement
- around 1/3rd of patients have a relapse



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HIV: renal involvement

Renal involvement in HIV patients may occur as a consequence of treatment or the virus itself. Protease inhibitors such as indinavir can precipitate intratubular crystal obstruction

HIV-associated nephropathy (HIVAN) accounts for up to 10% of end-stage renal failure cases in the United States. Antiretroviral therapy has been shown to alter the course of the disease.

There are five key features of HIVAN:

- massive proteinuria
- normal or large kidneys
- focal segmental glomerulosclerosis with focal or global capillary collapse on renal biopsy
- elevated urea and creatinine
- normotension

Hyperkalaemia: management

◆ Untreated hyperkalaemia may cause life-threatening arrhythmias. Precipitating factors should be addressed (e.g. acute renal failure) and aggravating drugs stopped (e.g. ACE inhibitors).

◆ Management may be categorised by the aims of treatment

Stabilisation of the cardiac membrane:

- intravenous calcium gluconate

Short-term shift in potassium from extracellular to intracellular fluid compartment

- combined insulin/dextrose infusion
- nebulised salbutamol

Removal of potassium from the body

- calcium resonium (orally or enema)
- loop diuretics
- dialysis

External Links

[BNF](#)

Management of hyperkalaemia

[The Renal Association](#)

Guidelines on the management of hyperkalaemia

Hypokalaemia and hypertension

For exams it is useful to be able to classify the causes of hypokalaemia in to those associated with hypertension, and those which are not

Hypokalaemia with hypertension

- Cushing's syndrome
- Conn's syndrome (primary hyperaldosteronism)
- Liddle's syndrome
- 11-beta hydroxylase deficiency*

Carbenoxolone, an anti-ulcer drug, and liquorice excess can potentially cause hypokalaemia associated with hypertension

Hypokalaemia without hypertension

- diuretics
- GI loss (e.g. Diarrhoea, vomiting)
- renal tubular acidosis (type 1 and 2**)
- Bartter's syndrome
- Gitelman syndrome

*21-hydroxylase deficiency, which accounts for 90% of congenital adrenal hyperplasia cases, is not associated with hypertension

**type 4 renal tubular acidosis is associated with hyperkalaemia

IgA nephropathy

Basics

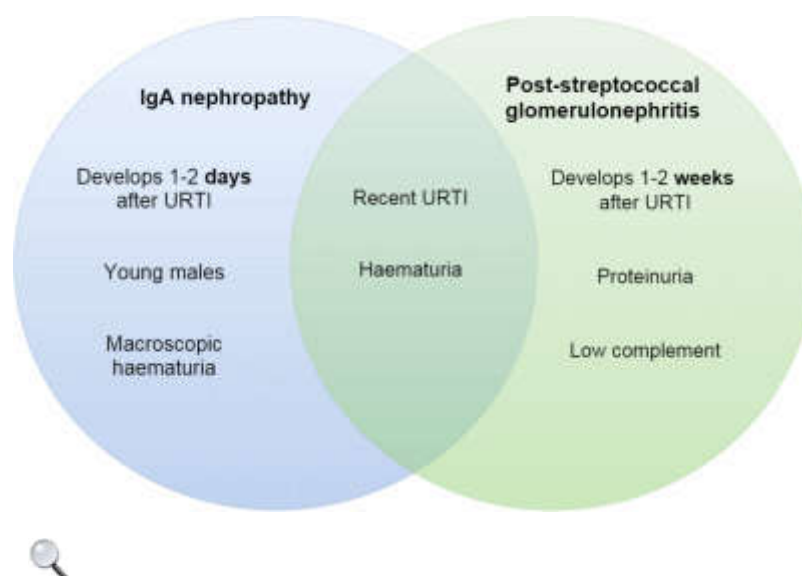
- also called Berger's disease or mesangioproliferative glomerulonephritis
- commonest cause of glomerulonephritis worldwide
- thought to be caused by mesangial deposition of IgA immune complexes
- there is considerable pathological overlap with Henoch-Schonlein purpura (HSP)
- histology: mesangial hypercellularity, positive immunofluorescence for IgA & C3

Presentations

- young male, recurrent episodes of macroscopic haematuria
- typically associated with mucosal infections e.g., URTI
- nephrotic range proteinuria is rare
- renal failure

Differentiating between IgA nephropathy and post-streptococcal glomerulonephritis

- post-streptococcal glomerulonephritis is associated with low complement levels
- main symptom in post-streptococcal glomerulonephritis is proteinuria (although haematuria can occur)
- there is typically an interval between URTI and the onset of renal problems in post-streptococcal glomerulonephritis



Associated conditions

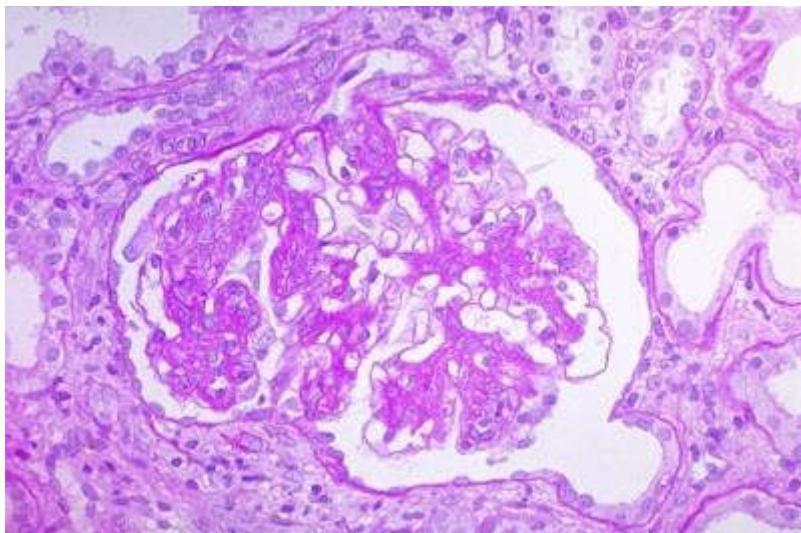
- alcoholic cirrhosis
- coeliac disease/dermatitis herpetiformis
- Henoch-Schonlein purpura

Management

- steroids/immunosuppressants not be shown to be useful

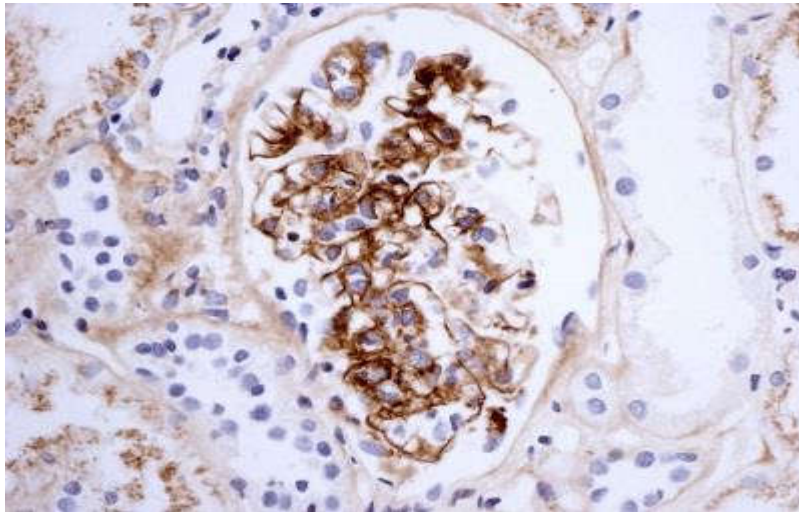
Prognosis

- 25% of patients develop ESRF
- markers of good prognosis: frank haematuria
- markers of poor prognosis: male gender, proteinuria (especially > 2 g/day), hypertension, smoking, hyperlipidaemia, ACE genotype DD



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Proliferation and hypercellularity of the mesangium is seen in the glomerulus



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Immunostaining for IgA in a patient with HSP

External Links

[Royal College of Physicians](#)

2012 Evaluation and management of IgA nephropathy

[Journal of Royal College of Physicians of Edinburgh](#)

What does STOP-IgAN tell us about how to treat IgA nephropathy?

Membranoproliferative glomerulonephritis

Overview

- also known as mesangiocapillary glomerulonephritis
- may present as nephrotic syndrome, haematuria or proteinuria
- poor prognosis

Type 1

- accounts for 90% of cases
- subendothelial immune deposits of electron dense material resulting in a 'tram-track' appearance
- cause: cryoglobulinaemia, hepatitis C.

Type 2 - 'dense deposit disease'

- causes: partial lipodystrophy, factor H deficiency
- reduced serum complement
- C3b nephritic factor (an antibody against C3bBb) found in 70%

Type 3

- causes: hepatitis B and C

Management

- steroids may be effective

Membranous glomerulonephritis

Membranous glomerulonephritis is the commonest type of glomerulonephritis in adults and is the third most common cause of end-stage renal failure (ESRF). It usually presents with nephrotic syndrome or proteinuria.

Renal biopsy demonstrates:

- electron microscopy: the basement membrane is thickened with subepithelial electron dense deposits. This creates a 'spike and dome' appearance

Causes

- idiopathic
- infections: hepatitis B, malaria, syphilis
- malignancy: lung cancer, lymphoma, leukaemia
- drugs: gold, penicillamine, NSAIDs
- autoimmune diseases: systemic lupus erythematosus (class V disease), thyroiditis, rheumatoid

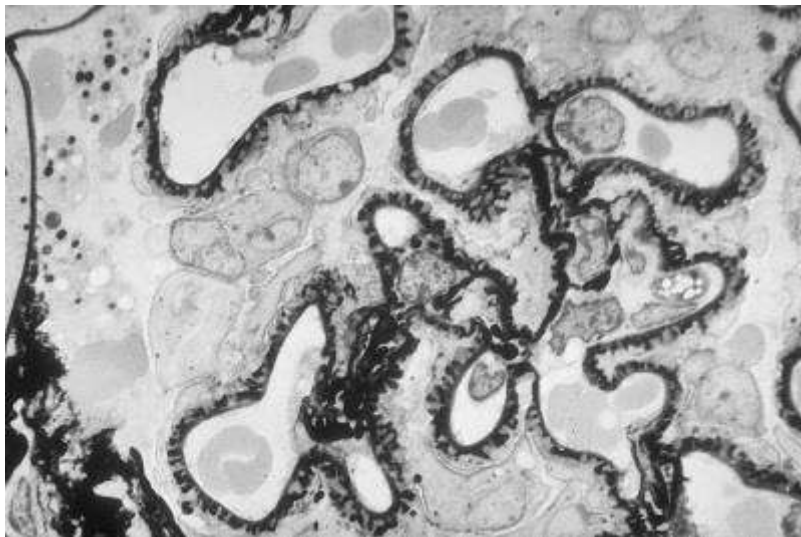
Prognosis - rule of thirds

- one-third: spontaneous remission
- one-third: remain proteinuric
- one-third: develop ESRF

Good prognostic features include female sex, young age at presentation and asymptomatic proteinuria of a modest degree at the time of presentation.

Management

- immunosuppression: corticosteroids alone have not been shown to be effective. A combination of corticosteroid + another agent such as chlorambucil is often used
- blood pressure control: ACE inhibitors have been shown to reduce proteinuria
- consider anticoagulation



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Silver-stained section showing thickened basement membrane, subepithelial spikes

External Links

[Royal College of Physicians](#)

2012 Membranous nephropathy review

Minimal change disease

Minimal change disease nearly always presents as nephrotic syndrome, accounting for 75% of cases in children and 25% in adults.

The majority of cases are idiopathic, but in around 10-20% a cause is found:

- drugs: NSAIDs, rifampicin
- Hodgkin's lymphoma, thymoma
- infectious mononucleosis

Pathophysiology

- T-cell and cytokine mediated damage to the glomerular basement membrane → polyanion loss
- the resultant reduction of electrostatic charge → increased glomerular permeability to serum albumin

Features

- nephrotic syndrome
- normotension - hypertension is rare
- highly selective proteinuria*
- renal biopsy: electron microscopy shows fusion of podocytes

Management

- majority of cases (80%) are steroid responsive
- cyclophosphamide is the next step for steroid resistant cases

Prognosis is overall good, although relapse is common. Roughly:

- 1/3 have just one episode
- 1/3 have infrequent relapses
- 1/3 have frequent relapses which stop before adulthood

*only intermediate-sized proteins such as albumin and transferrin leak through the glomerulus

Nephrotic syndrome

Triad of:

- 1. Proteinuria ($> 3\text{g}/24\text{hr}$) causing
- 2. Hypoalbuminaemia ($< 30\text{g/L}$) and
- 3. Oedema

Loss of antithrombin-III, proteins C and S and an associated rise in fibrinogen levels predispose to thrombosis. Loss of thyroxine-binding globulin lowers the total, but not free, thyroxine levels.

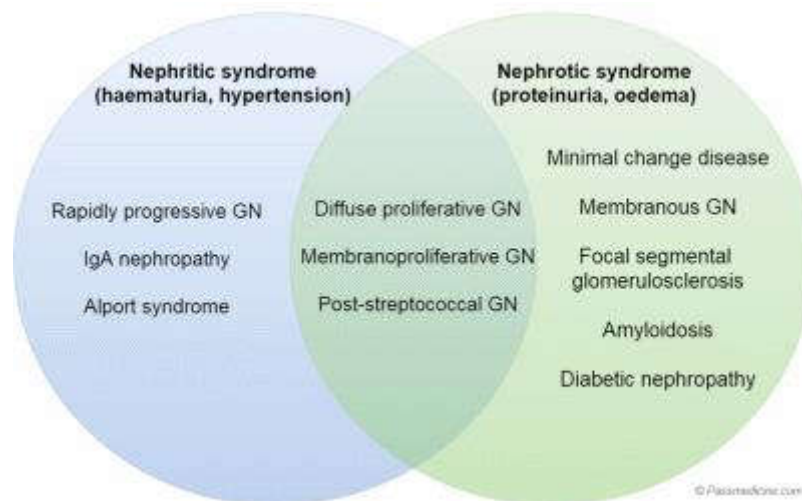


Diagram showing the glomerulonephritides and how they typically present

Nephrotic syndrome: causes

Primary glomerulonephritis accounts for around 80% of cases:

- minimal change glomerulonephritis (causes 80% in children, 30% in adults)
- membranous glomerulonephritis
- focal segmental glomerulosclerosis
- membranoproliferative glomerulonephritis.

Systemic disease (about 20%)

- diabetes mellitus
- systemic lupus erythematosus
- amyloidosis

Drugs

- gold (sodium aurothiomalate), penicillamine

Others

- congenital
- neoplasia: carcinoma, lymphoma, leukaemia, myeloma
- infection: bacterial endocarditis, hepatitis B, malaria

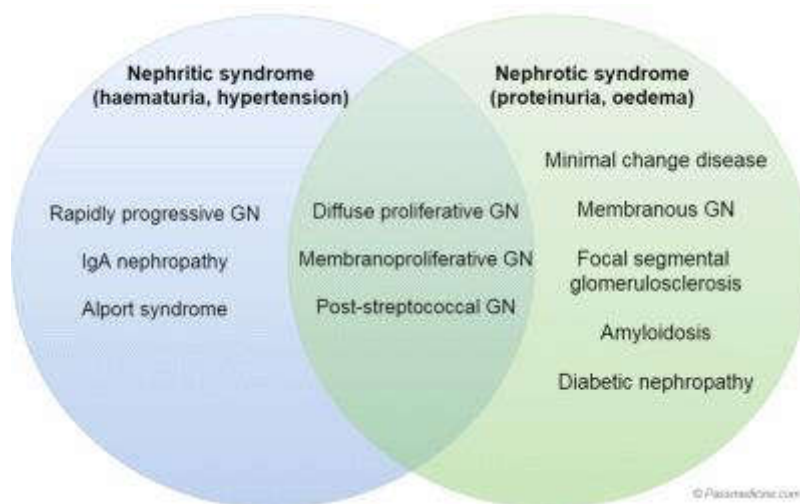


Diagram showing the glomerulonephritides and how they typically present

Nephrotic syndrome: complications

Complications

- increased risk of infection due to urinary immunoglobulin loss
 - increased risk of thromboembolism related to loss of antithrombin III and plasminogen in the urine
 - hyperlipidaemia
 - hypocalcaemia (vitamin D and binding protein lost in urine)
 - acute renal failure
-

Nephrotoxicity due to contrast media

Contrast media nephrotoxicity may be defined as a 25% increase in creatinine occurring within 3 days of the intravascular administration of contrast media.

Risk factors include:

- known renal impairment (especially diabetic nephropathy)
- age > 70 years
- dehydration
- cardiac failure
- the use of nephrotoxic drugs such as NSAIDs

Prevention

- the evidence base currently supports the use of intravenous 0.9% sodium chloride at a rate of 1 mL/kg/hour for 12 hours pre- and post- procedure. There is also evidence to support the use of isotonic sodium bicarbonate
 - N-acetylcysteine (usually given orally) has been shown to reduce the incidence of contrast-nephropathy in some studies but the evidence base is not as strong as for fluid therapy
-

External Links

[The Renal Association](#)

Guidelines relating to the prevention of acute kidney injury secondary to contrast

Papillary necrosis

Causes:

- chronic analgesia use
- sickle cell disease
- TB
- acute pyelonephritis
- diabetes mellitus

Features

- fever, loin pain, haematuria
- IVU - papillary necrosis with renal scarring - 'cup & spill'

Peritoneal dialysis

Peritoneal dialysis (PD) is a form of renal replacement therapy. It is sometimes used as a stop-gap to haemodialysis or for younger patients who do not want to have to visit hospital three times a week.

The majority of patients do Continuous Ambulatory Peritoneal Dialysis (CAPD), which involves four 2-litre exchanges/day.

Complications

- peritonitis: coagulase-negative staphylococci such as *Staphylococcus epidermidis* is the most common cause. *Staphylococcus aureus* is another common cause
- sclerosing peritonitis

External Links:

[Royal College of Physicians](#)

2012 End-stage renal disease review

Plasma exchange

Indications for plasma exchange (also known as plasmapheresis):

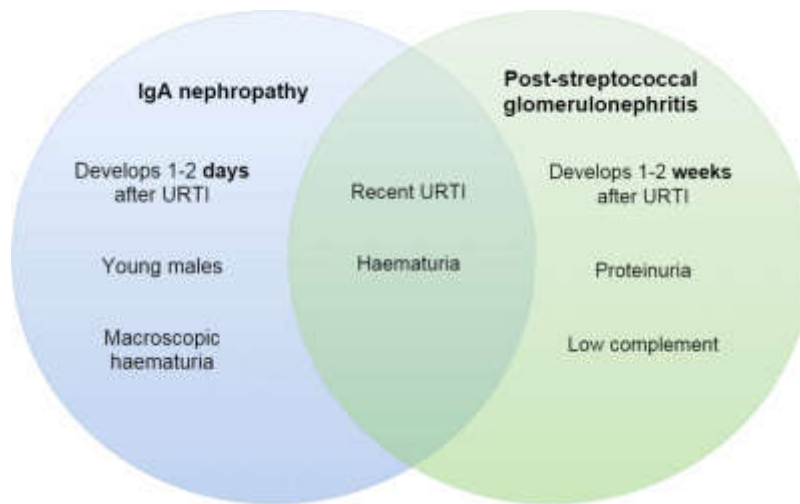
- Guillain-Barre syndrome
 - myasthenia gravis
 - Goodpasture's syndrome
 - ANCA positive vasculitis e.g. Wegener's, Churg-Strauss
 - TTP/HUS
 - cryoglobulinaemia
 - hyperviscosity syndrome e.g. secondary to myeloma
-

Post-streptococcal glomerulonephritis

Post-streptococcal glomerulonephritis typically occurs 7-14 days following a group A beta-haemolytic *Streptococcus* infection (usually *Streptococcus pyogenes*). It is caused by immune complex (IgG, IgM and C3) deposition in the glomeruli. Young children most commonly affected.

Features

- general: headache, malaise
- haematuria
- proteinuria
- hypertension
- low C3
- raised ASO titre

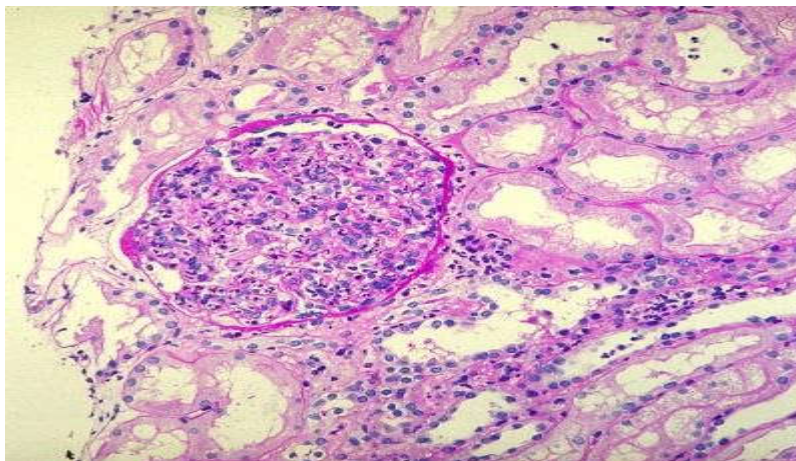


IgA nephropathy and post-streptococcal glomerulonephritis are often confused as they both can cause renal disease following an URTI

Renal biopsy features:

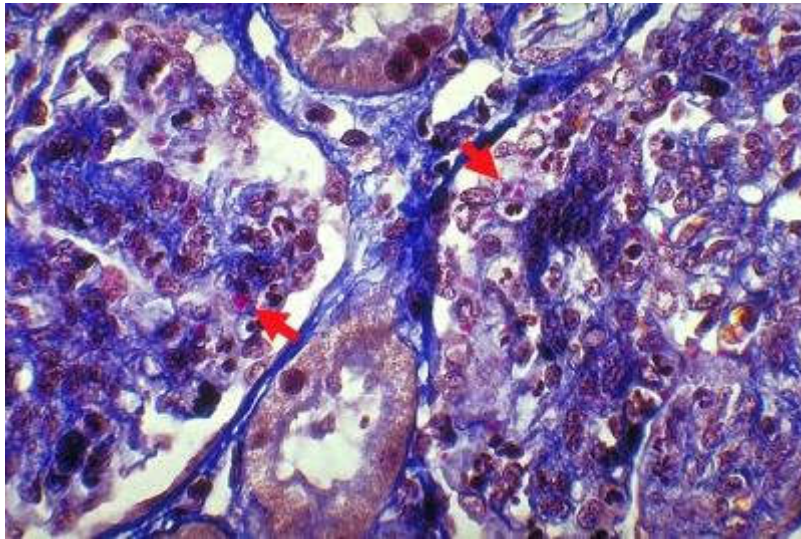
- post-streptococcal glomerulonephritis causes acute, diffuse proliferative glomerulonephritis
- endothelial proliferation with neutrophils
- electron microscopy: subepithelial 'humps' caused by lumpy immune complex deposits
- immunofluorescence: granular or 'starry sky' appearance

Carries a good prognosis



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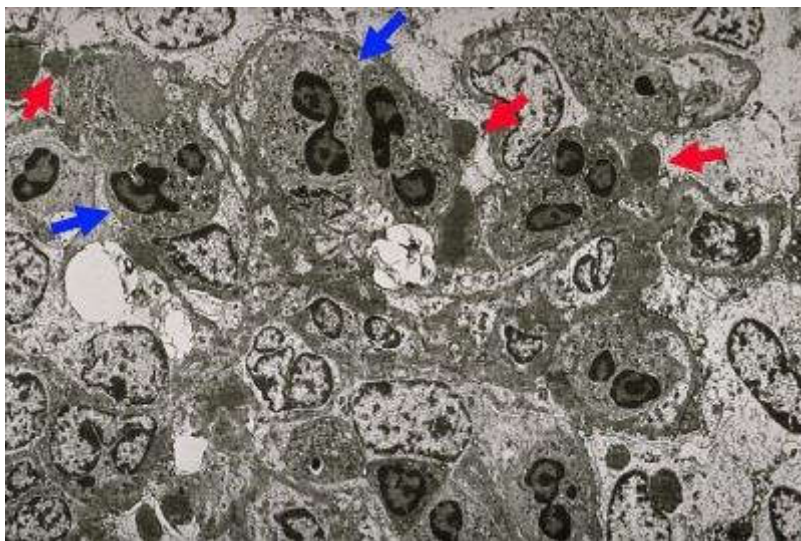
Proliferation of endothelium and mesangium with recruitment of neutrophils. Tubules are normal



© Image used on license from PathoPic



Subepithelial humps on the outside of the basal membrane



© Image used on license from PathoPic



Electron microscopy. Numerous neutrophils (blue arrows) and subepithelial humps (red arrows)

Prescribing in patients with renal failure

Questions regarding which drugs to avoid in renal failure are common

Drugs to **avoid** in renal failure

- antibiotics: tetracycline, nitrofurantoin
- NSAIDs
- lithium
- metformin

Drugs likely to accumulate in chronic kidney disease - **need dose adjustment**

- most antibiotics including penicillins, cephalosporins, vancomycin, gentamicin, streptomycin
- digoxin, atenolol
- methotrexate
- sulphonylureas
- furosemide
- opioids

Drugs relatively safe - **can sometimes use normal dose depending on the degree of chronic kidney disease**

- antibiotics: erythromycin, rifampicin
 - diazepam
 - warfarin
-

Primary hyperaldosteronism

Primary hyperaldosteronism was previously thought to be most commonly caused by an adrenal adenoma, termed Conn's syndrome. However, recent studies have shown that bilateral idiopathic adrenal hyperplasia is the cause in up to 70% of cases. Differentiating between the two is important as this determines treatment. Adrenal carcinoma is an extremely rare cause of primary hyperaldosteronism

Features

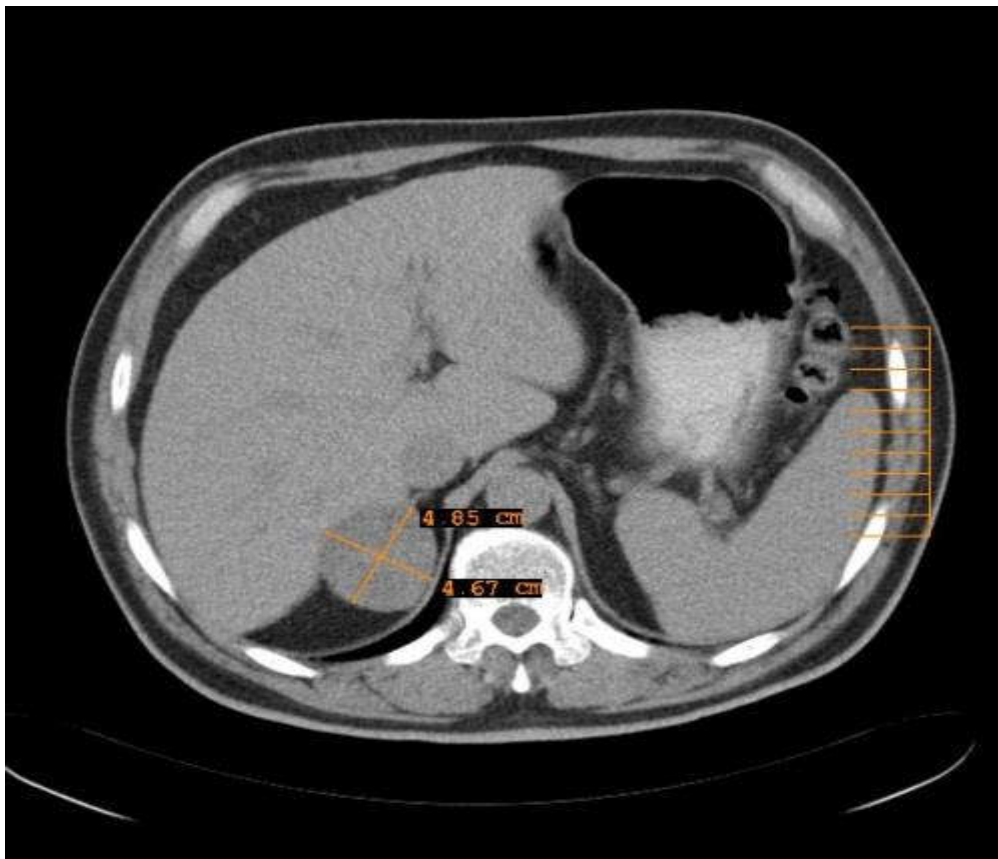
- hypertension
- hypokalaemia (e.g. muscle weakness)
- alkalosis

Investigations

- high serum aldosterone
- low serum renin
- high-resolution CT abdomen
- adrenal vein sampling

Management

- adrenal adenoma: surgery
- bilateral adrenocortical hyperplasia: aldosterone antagonist e.g. spironolactone



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CT abdomen showing a right-sided adrenal adenoma in a patient who presented with hypertension and hypokalaemia. The adenoma can be seen 'next to' or 'below' the liver.

Renal cell cancer

Renal cell cancer is also known as hypernephroma and accounts for 85% of primary renal neoplasms. It arises from proximal renal tubular epithelium

Associations*

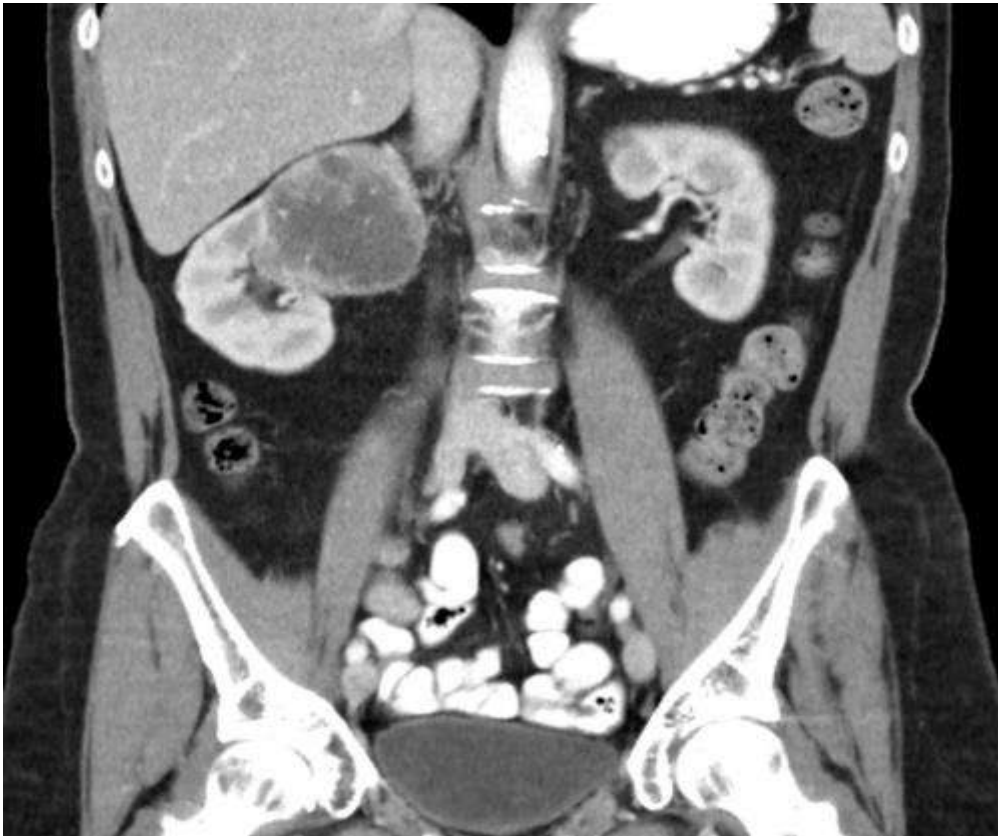
- more common in middle-aged men
- smoking
- von Hippel-Lindau syndrome
- tuberous sclerosis

Features

- classical triad: haematuria, loin pain, abdominal mass
- pyrexia of unknown origin
- left varicocele (due to occlusion of left testicular vein)
- endocrine effects: may secrete erythropoietin (polycythaemia), parathyroid hormone (hypercalcaemia), renin, ACTH
- 25% have metastases at presentation

Management

- for confined disease a partial or total nephrectomy depending on the tumour size
- alpha-interferon and interleukin-2 have been used to reduce tumour size and also treat patients with metastases
- receptor tyrosine kinase inhibitors (e.g. sorafenib, sunitinib) have been shown to have superior efficacy compared to interferon-alpha



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Coronal CT scan of a middle-aged woman with renal cell cancer. Note the heterogeneously enhancing mass at the upper pole of the right kidney

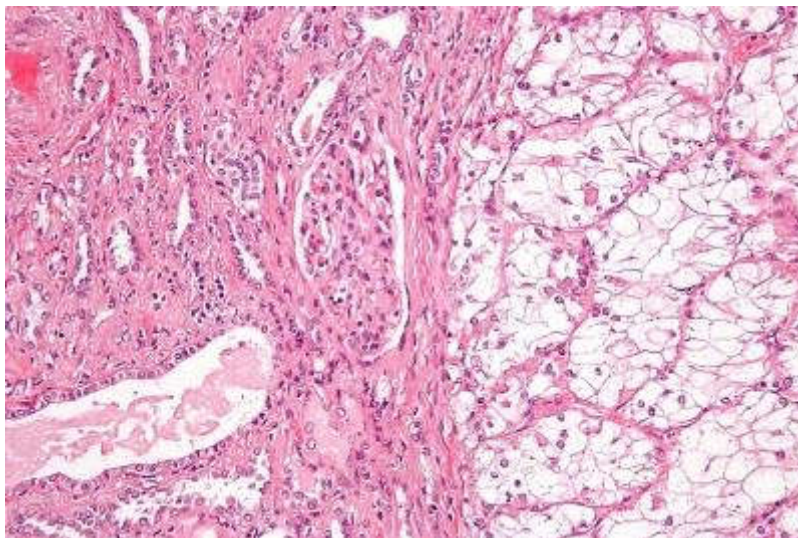
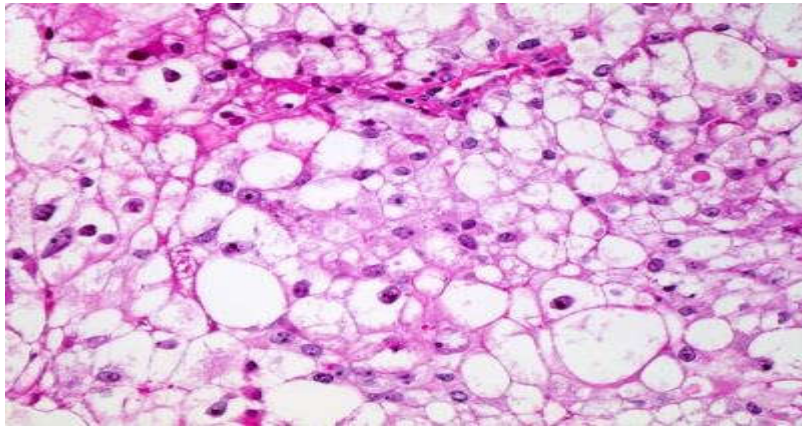


Image sourced from [Wikipedia](#) © Image used on license from PathoPic



Left: normal kidney. Right: 'clear-cell' pattern of renal cell carcinoma



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'Clear-cell' pattern of renal cell carcinoma - clear cytoplasm, small nuclei

*incidence of renal cell cancer is only slightly increased in patients with autosomal dominant polycystic kidney disease

Renal papillary necrosis

Renal papillary necrosis describes the coagulative necrosis of the renal papillae due to a variety of causes.

Features

- visible haematuria
- loin pain
- proteinuria

Causes

- severe acute pyelonephritis
- diabetic nephropathy
- obstructive nephropathy
- analgesic nephropathy: phenacetin was the classic cause but this has now been withdrawn
- sickle cell anaemia



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Renal papillary necrosis secondary to acute pyelonephritis. Note the pale areas highlighted by the blue arrows

Renal stones: imaging

The table below summarises the appearance of different types of renal stone on x-ray

Type	Frequency	Radiograph appearance
Calcium oxalate	40%	Opaque
Mixed calcium oxalate/phosphate stones	25%	Opaque
Triple phosphate stones*	10%	Opaque
Calcium phosphate	10%	Opaque
Urate stones	5-10%	Radio-lucent
Cystine stones	1%	Semi-opaque, 'ground-glass' appearance
Xanthine stones	<1%	Radio-lucent

*stag-horn calculi involve the renal pelvis and extend into at least 2 calyces. They develop in alkaline urine and are composed of struvite (ammonium magnesium phosphate, triple phosphate). *Ureaplasma urealyticum* and *Proteus* infections predispose to their formation

Renal stones: management

Initial management of renal colic

Medication

- the British Association of Urological Surgeons (BAUS) recommend diclofenac (intramuscular/oral) as the analgesia of choice for renal colic*
- BAUS also endorse the widespread use of alpha-adrenergic blockers to aid ureteric stone passage

Imaging

- BAUS guidelines recommend ultrasound as the initial imaging modality of choice. The sensitivity of ultrasound for stones is around 45% and specificity is around 90%. Complications such as hydronephrosis can also be quickly identified
- following an ultrasound, BAUS recommend a non-contrast CT (NCCT) to confirm the diagnosis. 99% of stones are identifiable on NCCT. Some GPs now have direct access to NCCT

Stones < 5 mm will usually pass spontaneously. Lithotripsy and nephrolithotomy may be for severe cases.

Management of renal stones

Most renal stones measuring less than 5mm in maximum diameter will typically pass within 4 weeks of symptom onset. More intensive and urgent treatment is indicated in the presence of ureteric obstruction, renal developmental abnormality such as horseshoe kidney and previous renal transplant. Ureteric obstruction due to stones together with infection is a surgical emergency and the system must be decompressed. Options include nephrostomy tube placement, insertion of ureteric catheters and ureteric stent placement.

In the non emergency setting the preferred options for treatment of stone disease include extra corporeal shock wave lithotripsy, percutaneous nephrolithotomy, ureteroscopy, open surgery remains an option for selected cases. However, minimally invasive options are the most popular first line treatment.

Shock wave lithotripsy

- A shock wave is generated external to the patient, internally cavitation bubbles and mechanical stress lead to stone fragmentation. The passage of shock waves can result in the development of solid organ injury. Fragmentation of larger stones may result in the development of ureteric obstruction. The procedure is uncomfortable for patients and analgesia is required during the procedure and afterwards.

Ureteroscopy

- A ureteroscope is passed retrograde through the ureter and into the renal pelvis. It is indicated in individuals (e.g. pregnant females) where lithotripsy is contraindicated and in complex stone disease. In most cases a stent is left in situ for 4 weeks after the procedure.

Percutaneous nephrolithotomy

- In this procedure access is gained to the renal collecting system. Once access is achieved, intra corporeal lithotripsy or stone fragmentation is performed and stone fragments removed.

Therapeutic selection:

Disease	Option
Stone burden of less than 2cm in aggregate	Lithotripsy
Stone burden of less than 2cm in pregnant females	Ureteroscopy
Complex renal calculi and staghorn calculi	Percutaneous nephrolithotomy
Ureteric calculi less than 5mm	Manage expectantly

Prevention of renal stones

Calcium stones may be due to hypercalciuria, which is found in up to 5-10% of the general population.

- high fluid intake
- low animal protein, low salt diet (a low calcium diet has not been shown to be superior to a normocalcaemic diet)
- thiazides diuretics (increase distal tubular calcium resorption)

Oxalate stones

- cholestyramine reduces urinary oxalate secretion
- pyridoxine reduces urinary oxalate secretion

Uric acid stones

- allopurinol
- urinary alkalinization e.g. oral bicarbonate

*Diclofenac use is now less common following the MHRA warnings about cardiovascular risk. It is therefore likely the guidelines will change soon to an alternative NSAID such as naproxen.

Chapter: Nephrology

External Links:

[European Urology Association](#)

2015 Urolithiasis guidelines

[Royal College of Physicians](#)

2012 Kidney stone disease: pathophysiology, investigation and medical treatment

[British Journal of Urology](#)

2015 Discussion regarding evidence base (or lack of it) for alpha blockers

Renal stones: risk factors

Risk factors:

- dehydration
- hypercalciuria, hyperparathyroidism, hypercalcaemia
- cystinuria
- high dietary oxalate
- renal tubular acidosis
- medullary sponge kidney, polycystic kidney disease
- beryllium or cadmium exposure

Risk factors for urate stones

- gout
- ileostomy: loss of bicarbonate and fluid results in acidic urine, causing the precipitation of uric acid

Drug causes:

- drugs that promote calcium stones: loop diuretics, steroids, acetazolamide, theophylline
- thiazides can prevent calcium stones (increase distal tubular calcium resorption)

Renal tubular acidosis

All three types of renal tubular acidosis (RTA) are associated with hyperchloraemic metabolic acidosis (normal anion gap)

Type 1 RTA (distal)

- inability to generate acid urine (secrete H^+) in distal tubule
- causes hypokalaemia
- complications include nephrocalcinosis and renal stones
- causes include idiopathic, RA, SLE, Sjogren's, amphotericin B toxicity, analgesic nephropathy



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Abdominal x-ray showing nephrocalcinosis - a classical finding in type 1 RTA

Type 2 RTA (proximal)

- decreased HCO₃⁻ reabsorption in proximal tubule
- causes hypokalaemia
- complications include osteomalacia
- causes include idiopathic, as part of Fanconi syndrome, Wilson's disease, cystinosis, outdated tetracyclines

Type 4 RTA (hyperkalaemic)

- reduction in aldosterone leads in turn to a reduction in proximal tubular ammonium excretion
- causes hyperkalaemia
- causes include hypoaldosteronism, diabetes

Renal vascular disease

Renal vascular disease is most commonly due to atherosclerosis (> 95% of patients). It is associated with risk factors such as smoking and hypertension that cause atheroma elsewhere in the body. It may present as hypertension, chronic renal failure or 'flash' pulmonary oedema. In younger patients however fibromuscular dysplasia (FMD) needs to be considered. FMD is more common in young women and characteristically has a 'string of beads' appearance on angiography. Patients respond well to balloon angioplasty

Investigation

- MR angiography is now the investigation of choice
 - CT angiography
 - conventional renal angiography is less commonly performed used nowadays, but may still have a role when planning surgery
-

Retroperitoneal fibrosis

Lower back/flank pain is the most common presenting feature. Fever and lower limb oedema is also seen in some patients.

Associations

- Riedel's thyroiditis
- previous radiotherapy
- sarcoidosis
- inflammatory abdominal aortic aneurysm
- drugs: methysergide

External Links:

[Patient.info](#)

Retroperitoneal fibrosis review

Rhabdomyolysis

Rhabdomyolysis will typically feature in the exam as a patient who has had a fall or prolonged epileptic seizure and is found to have acute renal failure on admission

Features

- acute renal failure with disproportionately raised creatinine
- elevated CK
- myoglobinuria
- hypocalcaemia (myoglobin binds calcium)
- elevated phosphate (released from myocytes)

Causes

- seizure
- collapse/coma (e.g. elderly patients collapses at home, found 8 hours later)
- ecstasy
- crush injury
- McArdle's syndrome
- drugs: statins

Management

- IV fluids to maintain good urine output
 - urinary alkalinization is sometimes used
-

Sterile pyuria

Causes

- partially treated UTI
 - urethritis e.g. *Chlamydia*
 - renal tuberculosis
 - renal stones
 - appendicitis
 - bladder/renal cell cancer
 - adult polycystic kidney disease
 - analgesic nephropathy
-

Thin basement membrane disease

An inherited disorder of type IV collagen that causes thinning of the basement membrane. It may affect up to 5% of the population and about 30% patients report a family history of haematuria. Diagnosis is usually based on the history of persistent haematuria, normal kidney function and family history of haematuria without kidney failure. It is generally a benign disorder and biopsy is rarely indicated.

Thrombotic thrombocytopenic purpura

Pathogenesis of thrombotic thrombocytopenic purpura (TTP):

- abnormally large and sticky multimers of von Willebrand's factor cause platelets to clump within vessels
- in TTP there is a deficiency of ADAMTS13 (a metalloprotease enzyme) which breakdowns large multimers of von Willebrand's factor
- overlaps with haemolytic uraemic syndrome (HUS)

Features

- rare, typically adult females
- fever
- fluctuating neuro signs (microemboli)
- microangiopathic haemolytic anaemia
- thrombocytopenia
- renal failure

Causes

- post-infection e.g. urinary, gastrointestinal
- pregnancy
- drugs: ciclosporin, oral contraceptive pill, penicillin, clopidogrel, aciclovir
- tumours
- SLE
- HIV

External Links

[British Committee for Standards in Haematology](#)

2003 TTP/HUS guidelines.

Wilms' tumour

Wilms' nephroblastoma is one of the most common childhood malignancies. It typically presents in children under 5 years of age, with a median age of 3 years old.

Features

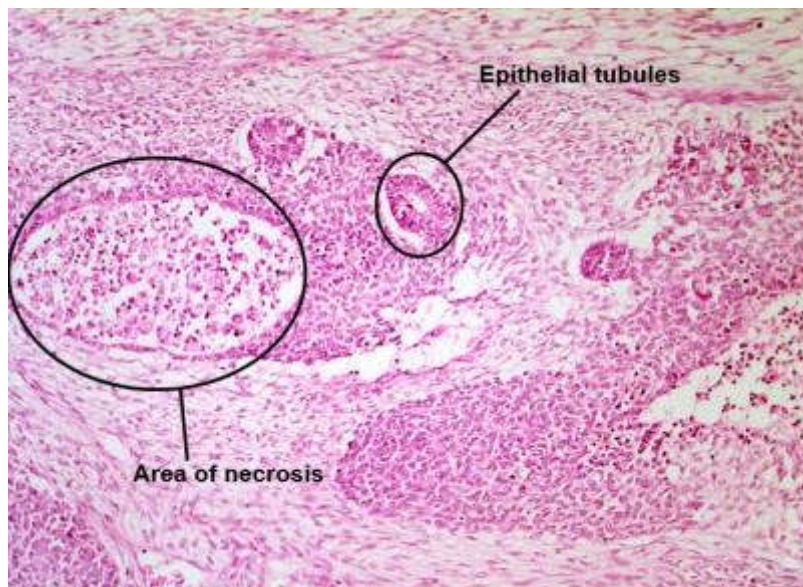
- abdominal mass (most common presenting feature)
- painless haematuria
- flank pain
- other features: anorexia, fever
- unilateral in 95% of cases
- metastases are found in 20% of patients (most commonly lung)

Associations

- Beckwith-Wiedemann syndrome
- as part of WAGR syndrome with Aniridia, Genitourinary malformations, mental Retardation
- hemihypertrophy
- around one-third of cases are associated with a loss-of-function mutation in the WT1 gene on chromosome 11

Management

- nephrectomy
- chemotherapy
- radiotherapy if advanced disease
- prognosis: good, 80% cure rate



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Histological features include epithelial tubules, areas of necrosis, immature glomerular structures, stroma with spindle cells and small cell blastomatous tissues resembling the metanephric blastema
